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CASE REPORT

Schwannoma of the base of tongue – A rare presentation

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KEYWORDS

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Abstract *Introduction:* Schwannomas are benign tumours arising from the Schwann cells. Only 1% of extracranial schwannomas occur intraorally and its pharyngeal presentations are rare.

Case report: We report a rare case of a 50 year old lady who presented with frequent hawking sensation since one year. Video laryngoscopy showed polypoidal lesion in the base of tongue. CT imaging was done to know the extent of lesion. Excision biopsy of the lesion was done. To our surprise, the histologic examination and the immunohistochemistry confirmation with S100 proved it to be schwannoma of the base of tongue.

Discussion: About a quarter of all schwannomas occur in the head and neck region. Only 1% of these occur intraorally, generally in the tongue. Pharyngeal presentations of schwannoma are rare.

Conclusion: Schwannomas arising in the base of tongue are rare and are not often included in the differential diagnosis. They are usually benign and have excellent prognosis as compared to the usual malignant lesions which occur in the tongue base.

Introduction

Neurilemomas or schwannomas, are usually solitary, slow growing benign neoplasm's that are well encapsulated soft tissue lesions deriving from Schwann cells. Schwannomas can arise from any cranial, peripheral, or autonomic nerves that contain Schwann cells, the cells that form the myelin sheath over nerve fibres.[1] Schwannoma was first reported by Verocay in 1910, who called this benign neurogenic tumour as neurinoma.[2]

About a quarter of all schwannomas occur in the head and neck region. Only 1% of these occur intraorally, generally in the tongue. Pharyngeal presentations of schwannoma are rare.[3] To the best of our knowledge only 13 cases of pharyngeal schwannomas have been reported and the tumour sites include posterior and lateral pharyngeal wall, pyriform sinus, and lateral glossoepiglottic fold.[4–9] Although they may arise at any age, the peak incidence of schwannoma is between the third and sixth decade of life, with no gender predilection.

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Case report

A 50 year old female presented with foreign body sensation and frequent hawking sensation in the throat for one year duration. There was neither history of difficulty or pain while swallowing or opening the mouth, nor history of change in voice, aspiration, nasal regurgitation or bleeding from the mouth.

On examination, the patient was of moderate build, with no pallor and the oral cavity examination was within normal limits. On depressing the tongue a mass was seen in the midline near the tongue base. Soft palate movements were normal. Indirect laryngoscopy showed a smooth surfaced pinkish polypoidal mass of 2×2 cm size attached to the base of tongue in the midline (Fig. 1). It was not extending into the valleculla. On palpation it was firm, non-tender, mildly indurated, with no bleeding to touch, nor attachment to surrounding tissue. The adjacent mucosa was normal. Mobility of the tongue was normal. No neck nodes were palpable. Video laryngoscopy confirmed the indirect laryngoscopy findings with a focal area of ulceration over the lesion. The rest of the oropharynx, hypopharynx and larynx were normal. Computed tomography scan showed mildly enhancing lobulated lesion attached to the base of tongue with no infiltration of adjacent tissue (Fig. 2).

The patient was prepared for trans-oral biopsy of the mass and was placed in Rose's position. Boyle Davis mouth gag with a shorter tongue blade was used for better visualisation of the tumour base and surrounding tissue. The tumour was seen to be attached to the tongue base by a pedicle and so we went ahead with an excision biopsy of the lesion and the base was cauterized. Postoperatively patient recovered well (Fig. 3).

Gross examination showed a mucosa covered pale yellow to white polypoidal nodule. Cut section showed smooth, pale white glistening tumour with streaks of haemorrhage (Fig. 4a). Histopathological examination showed a well-defined, circumscribed neural lesion covered by thinned out mucosa with ulceration. The tumour consisted of alternating hypocellular and hypercellular areas made up of spindle shaped Schwannian cells (Fig. 4b). The hypercellular (Antoni A) areas showed cellular interlacing fascicles of spindled cells with focal nuclear palisading around eosinophilic areas characteristic of Verocay bodies (Fig. 4c). The hypocellular (Antoni B) areas showed myxoid change. Blood vessels with thick hyalinised walls and focal thrombus formation were seen amidst the tumour. Immunohistochemistry showed strong S100 positivity in the spindle tumour cells (Fig. 5). Cytokeratin and SMA were negative in the tumour cells. Ki67 staining showed low proliferative index. Hence it was diagnosed to be a benign schwannoma arising in the base of tongue.

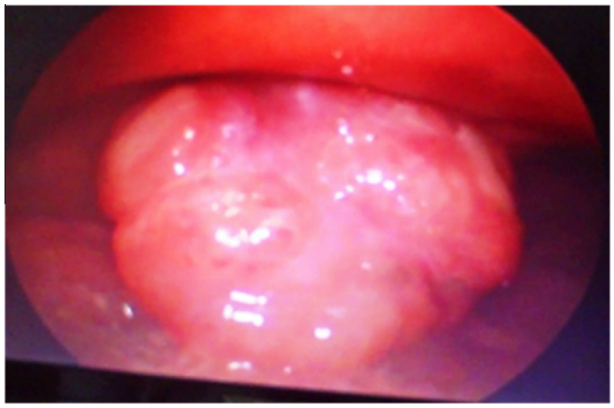

Figure 1 Tumour in the base of tongue as seen by video laryngoscopy.

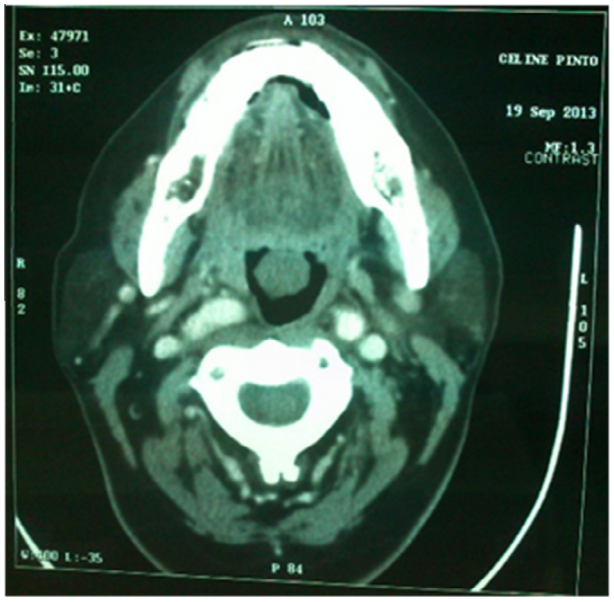

Figure 2 CT scan image showing mildly enhancing lesion attached to tongue base.

Discussion

The schwannoma is also called as neurilemmoma, neurinoma, perineural fibroblastoma and is a solitary, slow growing, usually encapsulated, asymptomatic tumour. The most common benign schwannomas encountered are acoustic neuromas occurring intracranially and affecting the eighth cranial nerve. Although a quarter of all extracranial schwannomas occur in head and neck, pharyngeal presentations are rare. So far, only about 13 cases have been reported in the base of tongue to the best of our knowledge.[4–9] It can present at any age, however it is more common between second and sixth decade of life.[1,2] They usually present with dysphagia, regurgitation, or snoring.[4–9]

The clinical differential diagnosis includes squamous cell carcinoma, soft tissue tumours and salivary gland tumours. Histologically, the differential diagnosis includes all spindle cell lesions at this site including sarcamatoid squamous cell carcinoma, smooth muscle, fibroblastic, lipomatous and rarely salivary gland tumours.[12]

In the present case the schwannoma presented as a slow growing, ulcero proliferative lesion in the base of tongue without any typical features of malignancy. Initially, schwannoma was not considered because of its rarity at this site. When a solitary polypoid lesion was found on table, an excisional biopsy was done.

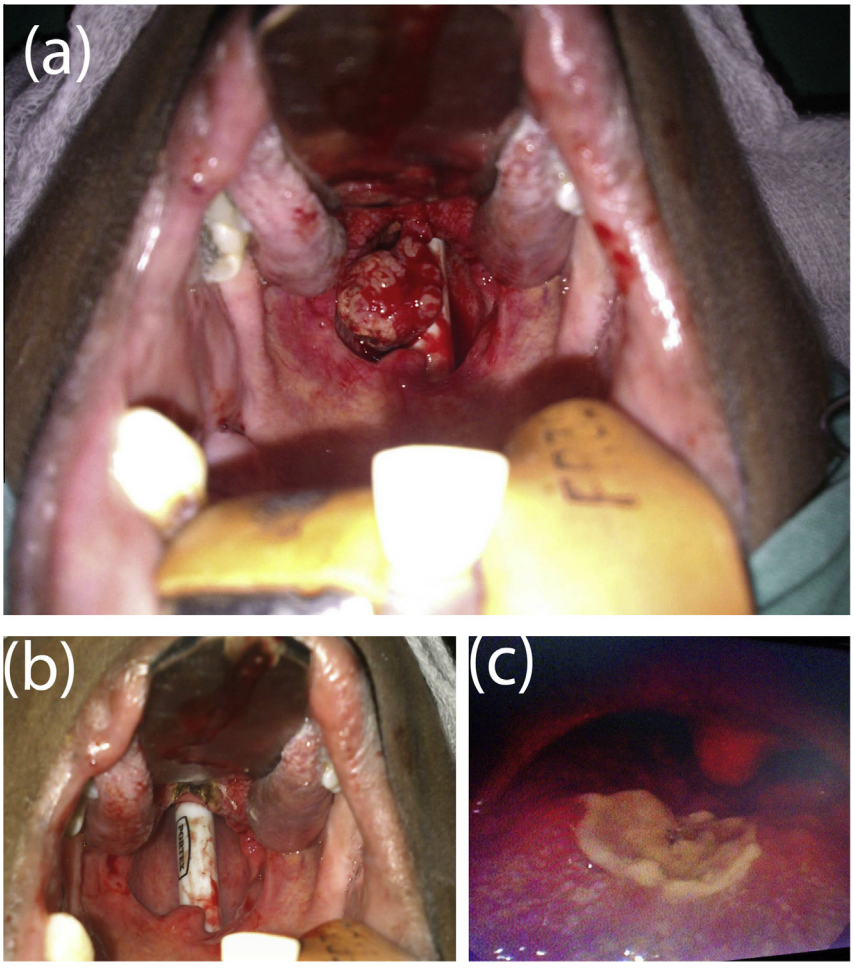

Figure 3 (Anticlockwise). (a). Pedicled tumour seen intraoperatively. (b). Post tumour excision and cautery. (c) 10 days follow-up.

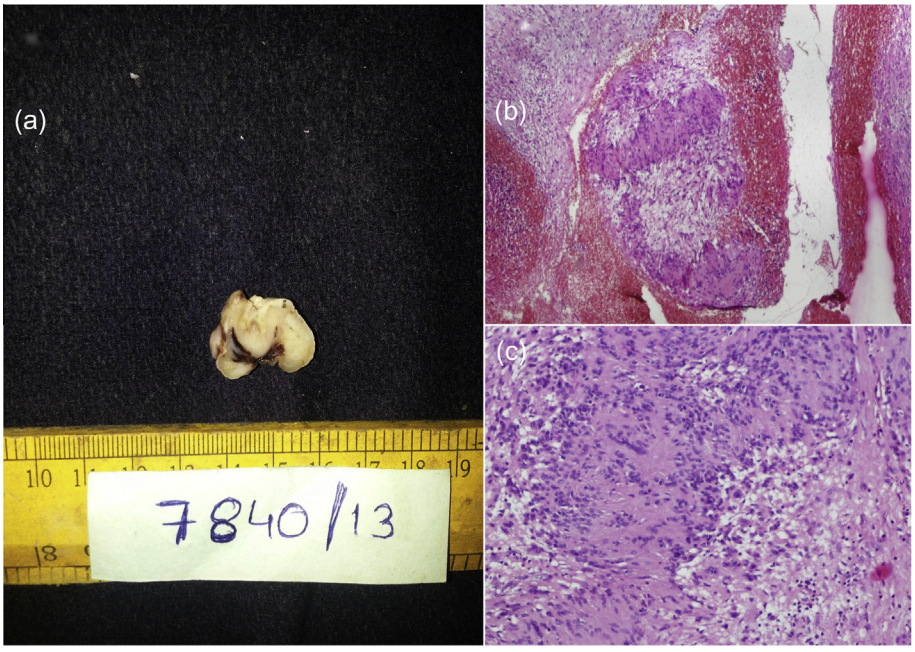

Figure 4 (Clockwise). (a) Gross section on cut surface showing pale white glistening surface. (b) Spindle cell tumours showing hypercellular and hypocellular areas (H&E, X100). (c) Verocay bodies seen in the hypercellular area (H&E, X400).

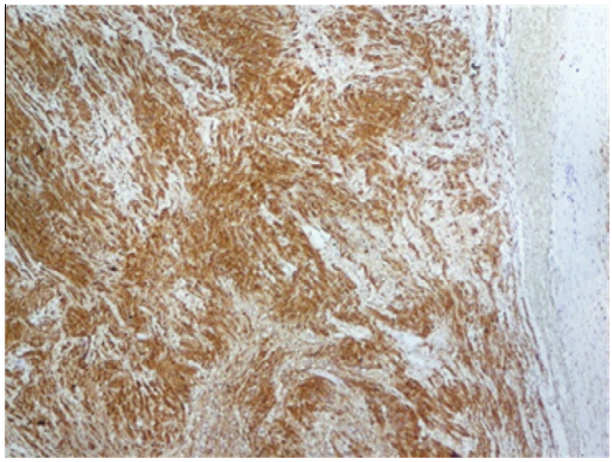

Figure 5 Immunohistochemistry for S100 protein showing strong positivity (×400).

Identification of the nerve of origin in these tumours may be difficult. In more than 50% of intraoral lesions, it is not possible to differentiate between tumours of the lingual, hypoglossal and glossopharyngeal nerves.[9] Schwannomas arising from the glossopharyngeal nerve are rare.[2]

Biopsy and use of imaging modalities are important in the preoperative diagnostic workup of any lesion of the head and neck.

Local excision or enucleation of these tumours is the treatment of choice.[10] The non-encapsulated form requires a margin of normal tissue and careful separation from the involved nerve is also necessary to preserve normal function. Schwannomas are not radiosensitive and so radiotherapy has no role in the treatment.[11] Recurrence is rare. Malignant transformation of a benign schwannoma is rare. In the present case connection with the nerve could not be seen, the mass was excised at the pedicle and base was cauterised. Prognosis is excellent as the tumour is benign, and recurrence is rare unless the resection of the tumour is incomplete.

We present this case for the rarity of schwannoma in the base of tongue, and to create awareness about immunohistochemistry to come to a definitive diagnosis.

Conclusion

The schwannoma of the base of tongue represents a lesion not often encountered in clinical practice. As a quarter of all extracranial schwannomas occur in head and neck, it should be considered as a differential diagnosis in benign lesions of oropharynx. The final diagnosis should be done after histopathological examination and in some cases after immunohistochemical analysis. The treatment of choice is total excision of the lesion and the prognosis is excellent.

Conflict of interest

None declared.

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